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Case report

Cutaneous plasmacytosis: Report of a Moroccan case!!

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Abstract

Cutaneous plasmacytosis is a rare entity that has been reported almost exclusively in Asian countries and is usually seen in adult males. Primary cutaneous plasmacytosis clinically is characterized by multiple red-brown plaques and nodules typically located on the trunk. We report a case of a Moroccan 65-year-old man presented multiple infiltrated red plaques on the extremities and the trunk, the diagnosis of cutaneous plasmacytosis was retained without systemic involvement. To our knowledge, this is the first case of this type reported in a Moroccan adult man.

Keywords: Cutaneous plasmacytosis; Systemic plasmacytosis; Lymphoplasmacytic disorder; Moroccan

1. Introduction

Cutaneous plasmacytosis is a rare skin disorder of unknown etiology. This condition has been recognized principally in Asian patients, with a few cases in whites (Xia et al., 2013). Patients presented clinically with multiple, red-brown infiltrated plaques and flat tumors, mainly located on the trunk. The condition can be systemic if polyclonal hypergammaglobulinemia, lymphadenopathy, cutaneous lesions, and systemic symptoms such as fever, weight loss, and fatigue are present (Cheng et al., 2012).

We report a case of 65-year-old man born and grew up in Rabat, presented a primary cutaneous plasmacytosis. To our knowledge, this is the first case of this type reported in a Moroccan adult man.

2. Case report

A 65-year-old male presented with a 2-year history of skin lesions that initially appeared on his lower limbs, then gradually thereafter on his trunk and upper limbs. The lesions were not pruritic. The patient was otherwise healthy and denied any systemic symptoms. His family history was unremarkable for skin disorders or lymphoproliferative disease. On physical examination, the patient had multiple 1 to 6 cm red-brownish macules and infiltrated plaques disseminated symmetrically on the trunk, abdomen, back, upper and lower extremities, sparing the face, scalp and palmoplantar regions (Figs. 1–4) with edema of the lower limbs. There was no enlargement of lymph nodes, liver or spleen.

A skin biopsy taken from his lower limbs showed the presence of a dermal nodular and perivascular mixed cell infiltrate with predominance of plasma cells without atyp-

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Fig. 1. Diffuse reddish-brown infiltrated plaques on the chest.

Fig. 2. Diffuse reddish-brown infiltrated plaques on the abdomen.

Fig. 3. Diffuse reddish-brown infiltrated plaques on the buttocks.

ical features. The epidermis was not involved (Fig. 5). Immunohistochemical study revealed polyclonality of plasma cells with expression of kappa and lambda light chain (Figs. 6 and 7 and 8).

Laboratory test results revealed no abnormalities in the hemogram. Tests for syphilis and HIV infection were negative. Serologic tests for hepatitis virus types B and C were negative. Serum protein electrophoresis detected polyclonal hypergammaglobulinemia with Ig G 35 g/l (normal, 7–16 g/l), Ig A 6.5 g/l (normal 0.7–4 g/l) and Ig M 3.78 g/l (normal 0.4–2.3). The urine electrophoresis study did not show Bence-Jones protein. A chest radiography and computed tomography scan revealed no apparent extracutaneous involvement.

Fig. 4. Diffuse reddish-brown infiltrated plaques on the upper limbs and lower limbs.

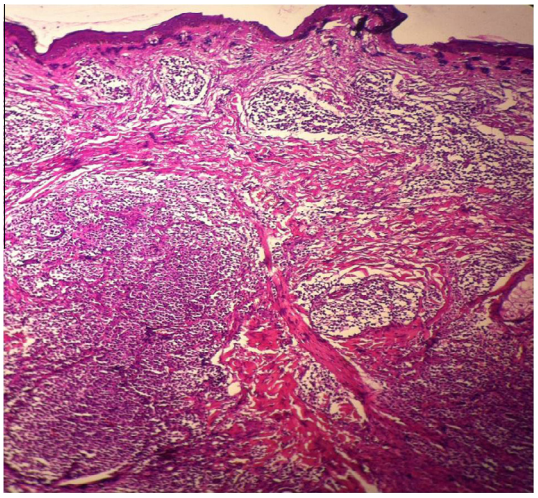

Fig. 5. Presence of dermal mixed cell infiltrate with predominance of plasma cells with perivascular and periadnexal infiltrates of plasma cells. Hematoxylin-eosin stain, original magnification: × 40.

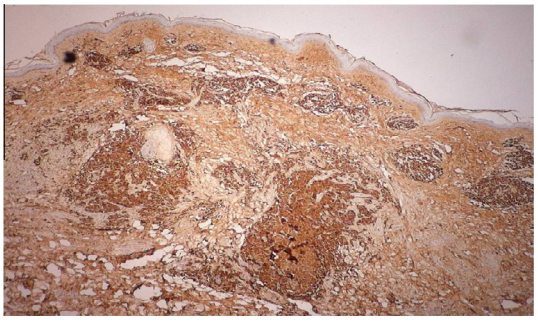

Fig. 6. Monoclonal expression of Kappa light chain by plasma cells.

Following the diagnosis of primary cutaneous plasmacytosis, oral corticosteroids were given with prednisone 40 mg/day which seemed to stabilize the lesions.

3. Discussion

Cutaneous and systemic plasmacytosis is a rare reactive lymphoplasmacytic disorder. It was first described in the

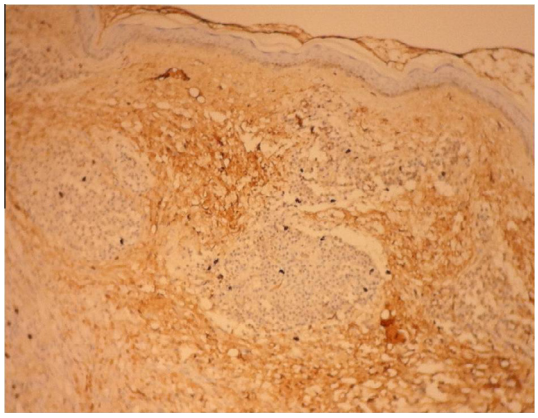

Fig. 7. Negative Lambda signal.

Japanese literature in 1976 by Yashiro as a kind of plasmacytosis and further characterized by Kitamura in 1980 (Honda et al., 2013; Leonard et al., 2007). Since that, it was described in Asian patients and particular in Japanese individuals (Shadel et al., 2010).

Cutaneous plasmacytosis is characterized by cutaneous lesions composed of mature polyclonal plasma cell infiltrates and polyclonal hypergammaglobulinemia (Uhara et al., 1994). The condition has been subsequently named cutaneous and systemic plasmacytosis when it is accompanied by anemia, fever, lymphadenopathy and, rarely, interstitial pneumonia (Shimizu et al., 1997).

Primary cutaneous plasmacytosis manifests as disseminated red brown cutaneous nodules or plaques mainly on the trunk, face or extremities (Ma et al., 2008). In our patient, the red indurated plaques on the extremities with edema of the lower limbs recalled the classic Kaposi's disease.

Histopathologically, lesions of cutaneous plasmacytosis are characterized by a dense perivascular infiltrates of mature plasma cells in the dermis without atypical features. The immunohistochemistry demonstrated the polyclonality of infiltrating plasma cells (Amin et al., 2002).

The etiology of cutaneous plasmacytosis is unknown, multiple theories have been proposed. The geographic distribution of cases has caused speculation that a primary infectious cause is responsible. The current evidence suggests that a degranulated production of interleukine 6 plays an important role in its pathogenisis (Kodama et al., 1992; Yamamoto et al., 1997).

Although the majority of patients with cutaneous plasmacytosis run a chronic benign course (Haque et al., 2011), a few cases have demonstrated an aggressive clinical course with a fatal outcome. There are reports of patients with plasmacytosis developing malignant lymphomas (Kanbe et al., 1998; Nitta, 1997).

It is essential to evaluate the patient for systemic disease and the development of a more aggressive disorder and the possibility of malignant transformation.

Available treatments for cutaneous plasmacytosis include topical and systemic corticosteroids, topical tacrolimus, systemic chemotherapy, topical photodynamic therapy, PUVA and radiotherapy with poor clinical responses (Miura et al., 2003; Tzung et al., 2005). In our patient, cutaneous lesions were stable after treatment with oral corticosteroids.

4. Conclusion

The dermatologist and dermatopathologist must be aware of this entity and its associations. It is essential to evaluate the patient for systemic disease and the development of a more aggressive disorder.

Description of new cases of cutaneous and systemic plasmacytosis in our country should prompt a search for infectious or environmental cause of the disease.

Conflict of interest

We have no conflict of interest to declare.

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